Leiomyosarcoma of the great saphenous vein

C. Werbrouck1, J. Marrannes1, P. Gellens2, B. Van Holsbeeck1, E. Laridon1

A 57-year-old man presented to his general practitioner with a palpable, painless mass in the right groin. There was no swelling of the ipsilateral leg. He was referred for diagnostic imaging. Ultrasound (US) identified a moderate circumscribed, heterogeneous hypoechoic mass in the right groin, probably in or next to the distal great saphenous vein, measuring approximately 18 x 25 x 26 mm. Power Doppler revealed a centrally strong vascular signal, with both arterial and venous signals (Fig. A). The origin of the mass was not entirely clear on US and subsequently an MRI and an US-guided puncture were performed. A homogeneous hypo-intense mass on T1 weighted imaging (WI) and slightly heterogeneous hyperintense lesion on T2 WI (Fig. B) was revealed, arising from the great saphenous vein, extending into the subcutaneous fat. The great saphenous vein was at least partially thrombosed, with a thickened wall and a mild surrounding infiltration. There were no susceptibility artefacts on gradient echo. Intravenous injection of gadolinium resulted in a strong enhancement in of the lesion (Fig. C, D). Small superficial veins ran into the lesion. The soft tissues surrounding the mass were edematous. There were multiple, slightly enlarged inguinal lymph nodes. No evidence for invasion of the adjacent inguinal canal or muscular structures was found. These finding were suggestive of a primary, possibly malignant tumor. Puncture of the lesion showed a spindle cell mesenchymal proliferation. Total resection of the lesion was performed (Fig. E, F), and anatomopathological examination showed a moderately differentiated intravascular leiomyosarcoma with limited pleomorphic dedifferentiation (5-10%). A subsequent CT of the thorax and the abdomen was negative for tumoral spread.

Comment

Leiomyosarcomas are aggressive soft tissue sarcomas arising from smooth muscle cells, and can be divided into three types according to their site of origin: soft tissue (most common), cutaneous (best prognosis) and vascular (arising from the muscular wall). Although five times more common than arterial leiomyosarcomas, primary venous leiomyosarcomas constitute for less than one in every 100,000 malignant tumours, and only 10% of these originate from the great saphenous vein. The inferior caval vein is the predominant venous location, accounting for almost half of the cases, followed by the pulmonary, renal, common femoral, saphenous, superior mesenteric, and ovarian veins, and superior caval vein. The development can be divided into three stages; nonocclusive stage (asymptomatic or non-specific symptoms), occlusive stage (ranging from asymptomatic to phlebitis or deep vein thrombosis), and terminal stage (distant metastases, especially in the lungs and liver). Color Doppler US is highly reliable in the early, non-occlusive stage. MRI and CT allow visualization of the tumor, determination of the venous origin, and relationship with surrounding structures. Current treatment of choice exists of limb-preserving surgery and adjuvant radiotherapy and/or chemotherapy (1).

Reference